

A CASE OF CEREBRAL HEMORRHAGIC PACHYMENINGITIS WITH PSEUDO-BULBAR PALSY.

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J. D., male, white, 43 years of age, a peddler, was admitted to the Philadelphia Hospital on April 29th, 1898, on account of epileptiform convulsions. His family history was negative. He had had acute articular rheumatism in 1891, and had used alcohol excessively for years. He denied venereal infection. His present trouble began in July, 1897, with a general convulsion after a debauch. He was unconscious and bit his tongue. Since the onset he has had at least one fit each week. On the day before his admission he had ten attacks, and during the previous week three. He has no distinct aura save momentary dizziness. He falls unconscious immediately. He usually passes a large quantity of urine after an attack. Headache lasting for several hours frequently follows. The attacks are increased in number and made more severe by drinking. For quite a long time he has had constant frontal headaches, at times so severe as to prevent sleep.

Condition on Admission.—He is a thin, pale, old-looking man. His gait is not ataxic, but he staggers a little, apparently from general weakness. There is no ataxia of station. He uses his hands well. Both knee-jerks are increased. Ankle-clonus is absent. The right grip is 45; the left, 32. There is no tremor of the hands, lips or tongue. Sensibility is normal over the entire body, head, trunk and extremities. He localizes touch well. Speech is slow and hesitating. During the first week in the hospital he had four convulsions with unconsciousness. After each fit he slept for about an hour. By May 15th he had become quite stupid, and at the same time very emotional, crying pitifully whenever spoken to. His gait became very uncertain, with a decided tendency to fall towards, or rather sink down on, the left side. He could not stand at

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all with the feet close together. The left grip was very poor, and the arm was held in a paretic attitude. The saliva dribbled. No convulsions occurred between the 9th and 23d. of May. The lower part of the face was then noticed to be drawn to the right, especially when crying. By May 24th the weakness on the left side had increased so much that he could not walk. He then began to pass urine and later feces involuntarily. This was due to his mental state, not to palsy of the bladder or rectum. He was dull, emotional, childish, demented. He understood little or nothing that was said, and his speech rapidly became inarticulate. By July 10th he had improved so much that he was able to be up and about the ward. On July 19th he had a convulsion, after which his speech again became bad, and he was once more helpless. During his stay in the hospital a few granular casts were occasionally found in the urine, but never albumin or sugar. He was discharged from the hospital greatly improved, September 6th, 1898, at the request of friends.

He was readmitted November 30th, 1898, complaining of weakness in both legs and difficulty in speaking.

Examination.—He is very poorly nourished. The left side of the face is relaxed, the left corner of the mouth drops, and the right is drawn upwards so that the mouth does not close perfectly on the left side. The tongue when protruded deviates to the left. It is not tremulous. There is great general emaciation, but no local wasting. The left arm is a little stiff. There is motor ataxia in both arms. When in bed he can lift either leg fairly well, and can flex and extend the toes easily, but movement at the ankles and knees is very weak. He cannot stand at all. The knee-jerks are very much increased. Passive lifting of the legs from the bed causes a clonus in them. Patellar clonus is marked on both sides. The legs are so spastic that ankle-clonus if present cannot be obtained. Both the biceps and triceps tendon jerks are present. Tapping the triceps muscle on the right arm causes clonus. The superficial reflexes are all present, and the plantar is especially marked. Sensibility is preserved over the entire body.

On examining the lungs, tactile fremitus is found increased over the right apex and upper lobe. The percussion note is not very unlike on the two sides. The breath sounds are harsh over both apices, especially the right. At the base posteriorly there are occasional coarse râles. The heart and abdominal organs are normal. The patient remained in the hospital only a few weeks, during which time he improved a great deal and was removed at the request of his friends. He was admitted for the third time on February 4, 1899.

Examination.—He is very much emaciated and very weak.

being bed-ridden. He coughs constantly, but cannot expectorate. He swallows with great difficulty. At first he could move his tongue freely, but after a few weeks it became almost immovable. Speech was as a rule inarticulate, but at times he could pronounce a word, clause, or even a short sentence distinctly. The left leg and arm and the lower half of the left side of the face were almost completely paralyzed. The arm and leg were contracted. He was very emotional, crying whenever spoken to, and indeed, whining and weeping for hours at a time. He seemed to understand all that was said to him. Both knee-jerks were much increased. Ankle-clonus could not be obtained, probably on account of the spasm. There were slight bedsores on both hips. Tactile sensibility was preserved. Pain and temperature sense were not tested. He paid no attention to the bladder or rectum.

Dr. de Schweinitz examined the eyes and reported: "Right eye: pupillary reaction normal. Left eye: old synechiæ from iritis. In right eye moderate optic neuritis, apparently of the edematous variety, the retina and nerve-head being cloudy with edema. Veins very full and between arteries normal in size. General absorption of the pigmented retinal epithelium. In left eye conditions similar, but not so marked."

The urine contained neither albumin nor casts.

He was weak mentally. He had quite a number of general convulsions. The temperature was, for the most part, normal, but there were several periods of fever up to  $103^{\circ}$ , lasting one, two and three days. He died April 29th, 1899.

On the following day a necropsy was held. The calvarium was normal. The dura was adherent to the brain over the entire convex portion of the prefrontal lobe on the left side, and over a circular area two and a half centimeters in diameter, at the apex of the right prefrontal lobe. The dura, where adherent, was very much thickened, and on cross section was seen to have an exudate about one-eighth of an inch in thickness on its outer, and a like exudate slightly thicker on its inner surface. On the inner surface of the dura a fibrinous pseudo-membrane covered the entire convexity on the left side and the frontal and parietal regions on the right. It stripped off easily, leaving the dural surface shining and hard.

The spinal dura from the upper cervical region to the level of the third thoracic segment showed a similar condition. Its greatest thickness was three-sixteenths of an inch. The delicate, fibrinous, pseudo-membrane could be traced to the fifth thoracic segment. The greatest thickening was on the ventral aspect of the cord, and gradually thinned out towards the dorsal aspect. Only the first and second cervical roots were caught in, and

compresed by, the thickened dura, the lower cervical and upper dorsal roots merely passing through, but not being injured by the delicate, soft pseudo-membrane.

Examination of the brain revealed on the second left frontal convolution several small partially caseated, partially organized tuberculous masses, varying in size from a barleycorn to a small cherry. These tubercles were in the cortex, penetrated the sub-cortical tissue slightly, and were situated beneath the dura at its thickest part. The rest of the brain showed no gross lesion. The convolutions were normal in size, the pia stripped readily, although it was somewhat thickened, and a careful microscopic examination of the brain substance, including the internal capsule, showed nothing to account for the motor disturbance. The crus, pons and medulla were very asymmetrical. The left side was distinctly smaller, the motor tract being one-half the size of the right, and lighter in color. Below the crossing of the pyramids, the spinal cord throughout its entire length was so much larger on the left side that we thought of a hemiatrophy secondary to the degeneration of the motor tract of that side. Inequality of the two sides is often seen in normal spinal cords, but cannot usually be traced into the opposite side of the bulb and peduncles. It is striking that the degenerative atrophy, visible to the naked eye, was on the side of the cord opposite to the hemiplegia. The microscopic examination explained this apparent anomaly. It showed the defect in the right side to be of long standing, whereas a recent acute degeneration was present upon the left. The patient had evidently had at some time a right hemiplegia, that had cleared up, leaving symptoms so few and insignificant that they were masked at the time of examination by the preponderance of those affecting the left side, and were considered to be due to general weakness. This was, perhaps, a pardonable error in judgment. That the lesion on the left side was not the so-called tertiary degeneration was proven by the clinical course of the case, and the presence of the lesion as high up as the internal capsule.

Microscopic examination of the central dura proved the external pachymeningitis to consist of two dense layers of newly formed fibrous connective tissue with oval and club-shaped nuclei, separated from each other and the true dura by areolar connective tissue, containing wide blood channels. The layers of the true dura were likewise thickened, with many enlarged and changing nuclei, but preserving perfectly their individuality from the newly formed membrane. The internal pachymeningitis followed the usual type, and presented a highly vascular newly formed fibrous layer, a layer of small, round cells and a mass of organizing fibrin on its free surface. In the ex-

ternal and internal layers evidence of old and recent hemorrhages from vessels with very thin walls was everywhere visible. The internal pachymeningitic membrane was very rich in blood vessels, which consisted mainly of a layer of endothelial cells lining fissures between the layers of fibrous connective tissue. The fibrinous layer contained many free red blood corpuscles and blood vessels in an early stage of formation in its mass. The layer of small, round cells with large, deeply staining nuclei appeared to result from a proliferation of the endothelium of the vessel walls on the free surface of the organized membrane. The marked thickening of the primary layers of the dura, the external pachymeningitis, the round cell infiltration between the layers of the internal pachymeningitis, all point to a true inflammatory process in the membranes of the brain. The condition of the tubercular masses in the cortex, their caseation and partial organization, and the condition of the neighboring cerebral tissue lead us to believe the following was the sequence of events:

An active tuberculosis of the cortex; a secondary inflammation of the dura from the irritation of the tubercular masses or by direct infection, the exudation of fibrin and its organization with the formation of a false membrane which followed the usual pathologic course. It is true that it was impossible to prove the presence of tubercle bacilli in the sections, but since the material was hardened in Müller's fluid, this was hardly to be expected. There were present in the round cell layer a number of bacilli which were of the size and shape of tubercle bacilli, but they did not stain characteristically. Further the tissue examined showed the structure usually found in the false membrane of hemorrhagic pachymeningitis, and was not at all tuberculous. The tubercles of the cortex were typical in structure, and particles of these injected into rabbits gave positive results.

Microscopical examination of the cerebral cortex revealed the following: The usual carmine and nuclear stains stained the tissue as one diffuse mass, the cells and vessels being distinguished only by their pericellular and perivascular spaces. This total lack of differentiation depends, we think, on a chemical change in the tissue, resembling the albumose gelatin exudate described by Echeverria. By staining first in ammonium carmine, then by fuchsin, and overstaining with hemalum the following changes are noted: The subpial layer is very much thickened. There is a diffuse gliar sclerosis characterized by an increase in the number of the gliar cells, with hypertrophy and fat infiltration and a thickening of the gliar network. The smaller ganglion cells have large vesicular

nuclei, their apices point in all directions; a leucocytic infiltration is seen in some and a pathologic pigmentation in all. As to this last condition, though the amount of pigment varies normally, it never in health occupies the entire cell body as was the case here in many cells, or even two-thirds, or one-half



Fig. 1. Section of cortex, motor area. Marchi's stain. The Deiters' cells in the subpial layer show black dots of fat. The hypertrophied glia cells in the white matter are indicated by the accumulations of black dots in the lowermost section of the sketch.

of the cell area, as happened in all. Abnormal pigmentation, however, does not necessarily mean disturbance of function. This pigment staining black with osmic acid was very striking

and extensive. Another pigment staining black with hematoxylin was present in most of the ganglion and glia cells. It is evidently a distinct pigment and of doubtful origin. This double pigmentation was not confined to the cortical ganglion cells, but was found throughout the entire central nervous sys-

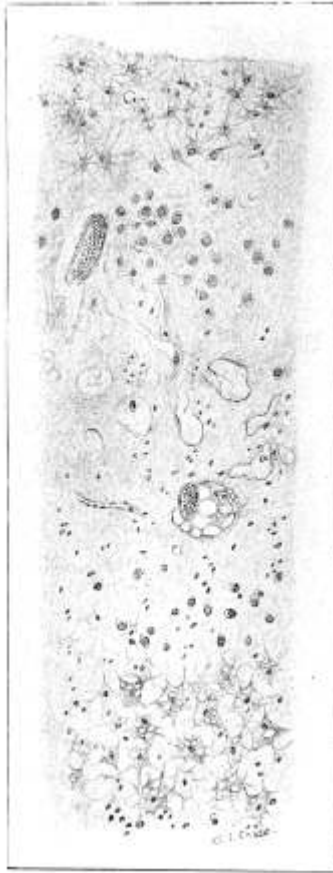


Fig. 11. Section of cortex, motor area. Rosin's stain. The hypertrophy of the subpial glia cells, and of the glia cells in the subcortical white matter, is shown. The cerebral porosis, with a glia cell in one of the spaces near the middle of the picture, is indicated; as is also the scarcity of large pyramidal cells.

tem. The large pyramidal ganglion cells in the deeper layers of the cortex are few in number and show the same changes noted in the smaller pyramidal cells. The so-called leucocytes

in the pericellular spaces and apparently at times in the nerve cells when stained by Ehrlich's triple stain resemble the lymphocytes of the blood.

The tangential fibers in the cortex were fewer than normal, and widely separated in the thickened subpial layer. The terminal projection fibers were also decreased in number, and more widely separated than normal. Association fibers were not observed anywhere in the sections. The gliar meshwork in the entire cortex was much closer and coarser than normal, except in the neighborhood of the blood vessels, where a rarefaction had taken place with the formation of microscopic vacuoles, within which gliar cells and gliar fibers could be observed. (See Figs. I and II.) This is the cerebral porosis of Marinesco and Sérioux. That it is not, as many maintain, simply a dilatation of the perivascular spaces is shown by the presence of glia cells with spider-like processes within the spaces, by the normal caliber of the perivascular spaces of vessels in the immediate neighborhood, and by the peculiar eccentric arrangement of the vacuoles, in many cases extending out from one side of the vessels only. The absence of free red blood corpuscles in the tissue speaks against any marked congestion with dilatation of the capillaries. The whole appearance suggested the absorption of cerebral substance in the presence of a diffuse hypertrophy of the glia—an early stage of a process which might end either in a condition of multiple macroscopic vacuolization of the cortex (Stühlinger and Fürstner), or, as seems more probable, a sclerotic atrophy of the affected convolutions. Examination of the underlying white matter supports this view. The gliar cells formed there a close meshwork, were much hypertrophied, at times four or five times their normal size, and showed in the cell body large fatty granules. This condition probably depends on an absorption by the glia cells of the fatty detritus of the breaking down cerebral substance.

The cerebellum presented the same peculiar staining properties, the same hypertrophy with fat infiltration of the glia, and a rarefaction of the cerebellar substance, modified by the peculiarities of its structure. By the Marchi method, a continuous layer of glia cells, infiltrated with fat, was found at the outer margin of the granular layer. By other stains, a distinct rarefaction in the immediate neighborhood of this layer was shown, presenting the appearance of lacework. In the pons and medulla the same fat infiltration of the cells was seen.

The cells of the spinal cord presented the same double pigmentation above referred to. Instead of the atrophy noted in the cells of the cortex, the cells of the anterior horns were,

if anything, larger than normal. This may have been due to the large accumulation of pigment in the cells. The cell nuclei were centrally located, the processes seemed normal and stained well.

In the white matter Weigert's hematoxylin method showed descending degeneration very marked in the direct pyramidal tract of the left side and the crossed pyramidal tract of the right. It also showed a slight degeneration in the opposite motor tracts. Marchi's method reversed the picture, that is, the most marked degeneration was found on the left side which showed but little change with the Weigert method; the right direct pyramidal tract was also much degenerated and could be traced into the sacral cord. Slight degeneration was shown by the Marchi method in the right crossed pyramidal tract and the left direct tract. In the cervical region a distinct degeneration was noted in Loewenthal's tract. The tracing of these fibers by Marchi's method in and above the bulb had to be omitted, in order to determine whether the case was one of cerebro-bulbar or pseudo-bulbar palsy. The presence of the membrane in the cervical region may have had something to do with the degeneration of Loewenthal's tract. In the upper cervical cord the posterior columns were degenerated, and sections of the posterior roots where they passed through the thickened dura showed the latter to be the cause of the degeneration. In the thoracic and lumbar regions the posterior columns did not show degeneration by the Marchi method; but the carmine and Weigert hematoxylin stain showed sclerosis in Goll's columns. There was marked sclerosis in the left direct and right crossed pyramidal tracts, and a slight sclerosis in the opposite motor tracts.

Macroscopic examination of the thoracic and abdominal viscera by Dr. D. Riesman revealed old and recent tuberculosis, and bilateral gangrene of the lungs; chronic interstitial nephritis; beginning aortic atheroma.

The discovery of the primary cause of the various pathological conditions narrated above resolves itself into a study of the etiology of the hemorrhagic pachymeningitis. Several factors must be considered. Foremost among them is the continued and excessive use of alcohol. It is pretty well proven that it alone may be a sufficient cause. It is not proven, however, that it is always the cause. We have seen more than once a tumor of the cortex, associated with a widespread pachymeningitis, having no other apparent cause than the tumor

itself. The influence of tuberculosis in the development of the condition is of no little importance. That some relation exists between the two diseases has been frequently noted. Of forty-one cases reported by Wiggelsworth, most of which were in the insane, twenty-two had general paralysis, and nine phthisis. In six cases of phthisical insanity reported by Dercum marked changes in the meninges were noted. Fürstner, after calling attention to the presence of delicate pseudo-membranes on the dura of consumptives, says he has so often seen in drunkards, immediately after the first signs of pneumonia, brain symptoms caused by a hemorrhagic pachymeningitis that he is sure the latter was produced by the circulatory disturbance resulting from the infiltration of extensive lung areas. Barthéz and Sanné differentiate intracranial hemorrhage in tuberculous children into medullary hemorrhage more common in acute miliary tuberculosis, and meningeal hemorrhage when the tuberculosis is of the chronic type with glandular enlargements and marked cachexia. Goldscheider and Raymond have reported cases of circumscribed, chronic, tuberculous meningitis of the spinal dura. Indeed a mere glance at the literature shows the frequent association of the two conditions, but whether there is a causal relation between them or not is an unsettled question. There is no reason why the fibroid changes noted in the pleural, pericardial and peritoneal cavities in phthisis should not take place in the intrameningeal spaces. We have already stated the causal relation we believe to have existed between the tuberculous masses of the cortex and the false membrane and dural thickening.

As to the spinal pachymeningitis, in the absence of any local tuberculous growth, it is probable that alcohol was the primary cause.

The diagnosis of hemorrhagic pachymeningitis is always difficult, frequently impossible. It occurs most often in insanity which masks its symptoms, or it is, as in our case, one of many lesions. It may exist for years without appreciable symptoms. It may simulate a variety of conditions, as tuberculous meningitis, progressive paralysis, and brain tumor. Occasionally, however, there is a certain symptom-complex, a particular clinical course from which we may more than guess its

presence. Thus given a man who has used alcohol for years, who is syphilitic or in whom senile changes are marked, the occurrence of an apoplectic or epileptic attack, especially if preceded for some time by headache and vertigo and not presenting symptoms of brain tumor, at least creates a suspicion of hemorrhagic pachymeningitis. The coma of the acute attack is usually not so deep as that in hemorrhage within the brain. There are always symptoms of irritation, contracted pupils, delirium, monoplegic or hemiplegic rigidity. The palsy is not primarily flaccid, as in intracerebral hemorrhage, but spastic from the first.

In days or weeks the patient recovers in greater or less degree mental and physical power, only to lose them again by a second hemorrhage from the partially organized membrane. The attacks recur, and after each the patient is left more palsied and with less mentality, until finally he dies from an apoplectic seizure or from some complicating disease of the lungs or kidneys. The rigidity and increased reflexes at the onset, the absence of cranial nerve involvement, and above all the marked remissions, are important in diagnosis. In cases occurring in the chronic insane in which the hemorrhage is not sufficient to fill up the space caused by the wasting of the convolutions, there often are no noticeable symptoms.

Our patient after a life of drunkenness developed when about forty years old a disease supposed by the committing physician to be late epilepsy. While under observation he began to suffer from intense headache, vomiting, left hemiparesis going on to almost complete palsy, failure of memory and great loss of mental power, and a marked increase in the number of convulsions—a clinical picture which led to the tentative diagnosis of brain tumor. When, however, the exacerbations and remissions became more marked, a slowly forming optic neuritis appeared, palsy of the faucial muscles, ataxia of the arms, difficulty in swallowing, in expectoration, in articulation and in the movements of the tongue, and finally the so-called forced emotional states, so frequently seen in bulbar disease, the difficulties of diagnosis were greatly increased.

Bulbar disturbances are not infrequent in the last stages of

organic insanities, but in this case the ordinary sequence of events was reversed, the dementia following instead of preceding the appearance of the symptoms of gross organic lesions. In form our diagnosis, brain tumor, was correct—tumors were found post mortem, but in fact it was an error, for the whole picture seen at autopsy was unlike that conceived by us while the patient was alive. The exacerbations and remissions were consistent with hemorrhagic pachymeningitis. Inflammatory changes in the optic nerves have also been observed in hemorrhagic pachymeningitis, but not of the kind present in this case. As shown by Fürstner, Griesinger and Oppenheim, there may be an acutely developing choked disk, due as proven by necropsy to an infiltration of blood into the optic sheath. In our case the development was slow, and shortly before death only a marked, cloudy edema of the nerve and retina was observed, probably due to slowly developing and long-continued increased intracranial pressure.

We were thrown off the proper diagnostic track also by the cranial nerve involvement. According to Bullard and others this is very rare in hemorrhagic pachymeningitis. When it does occur the necropsy has always revealed a hemorrhagic infiltration about the base of the brain. This was not present in our case, and it is of interest to determine what was the causation of the bulbar symptoms. That multiple foci of disease in the cerebrum may, though rarely, give rise to symptoms closely resembling the Duchenne type of progressive bulbar paralysis, without any lesion in the bulb, is well established, and that this is the explanation of our case is proven by the microscopic examination. Since serial sections of the bulb show no areas of inflammation or degeneration, we have concluded that a true cerebral form of bulbar paralysis was present, and not the cerebro-bulbar form, in which lesions are found both in cerebrum and medulla.

The cervical pachymeningitis had of course nothing to do with the bulbar symptoms, and caused only the ataxia in the arms. The clinical course was more that of the true cerebral type—a slow and gradual increase in difficulty in swallowing,

progressive slurring of speech until speech became a mere jumble of noises with now and then an explosion into some short commonplace sentence. The forced and spasmodic emotionalism is common in this syndrome, and was very marked in this patient. He would attempt to talk and after a few efforts suddenly break down and begin to cry. Whether this emotion was real, or whether it was only a reflex mimicry of grief, must remain unsolved. This case furnishes a striking example of the pseudo-bulbar syndrome resulting from a purely cortical condition. Pachymeningitis must be a very rare cause. The tubercles, we think, may be left out of consideration as causative factors, because of their position, their size, their long duration before bulbar symptoms appeared, and the fact that they were all in one hemisphere. This last fact is important. The cases of Magnus, Bamberger, Kirchoff, etc., in which unilateral lesions are described as having produced bulbar symptoms, cannot be accepted as accurate in the light of newer methods of microscopic research. In the absence of a thorough microscopic examination, the probability is always in favor of the cerebro-bulbar type of the disease. For a unilateral cerebral lesion to cause the condition would require an anomalous distribution of cortico-bulbar innervation.

The diffuse cortical sclerosis is a possible causative factor, but there is no reason to suppose that in the absence of the pachymeningitis, it would have produced bulbar symptoms. Jolly's case of pseudo-bulbar paralysis with multiple cerebral sclerosis did not resemble ours, clinically or pathologically. In it there were well defined sclerotic patches cutting off certain brain tracts.

The following conclusions may be drawn:

1. Hemorrhagic pachymeningitis should not be forgotten as a possible cause of epileptiform fits beginning in adults.
2. The condition may sometimes at least be inflammatory in origin.
3. Alcohol and tuberculosis are both causes.
4. A pseudo-bulbar palsy may develop in the course of the disease.